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*Some Observations on the Hereditary Form
of Chorea, with the Report of a Case.*

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SOME OBSERVATIONS ON THE HEREDITARY FORM OF
CHOREA, WITH THE REPORT OF A CASE.

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VON ZIEMSEN,¹ in his comprehensive and exhaustive treatise on chorea, says, under the head of etiology: "Hereditary transmission does occur, but is certainly rare; but hereditary *tendency* to chorea, as to other diseases of the nervous system, . . . is very often demonstrable." He makes no further reference to the subject, except to quote Huntingdon's remarkable report,² in which there are related the records of several families living on Long Island, nearly all of the members of which were affected with chorea. Huntingdon and his father and grandfather together were able to trace a relationship between these families, and a direct hereditary transmission of the disease through several generations. In these cases the disease began between the ages of twenty and forty, attacking men and women alike. If a fortunate brother or sister of these choreics escaped the affliction, his or her children were also exempt.

W. P. Herringham,³ in an admirable critical digest on "Chorea in the Adult and in the Old," says: "Chorea is not a common disease in persons after thirty years of age, and the records of it are scanty; but the term is used of so many different classes of illness, that it is worth while to arrange and classify the cases at present recorded." He makes four classes, or divisions, as follows:

First, chorea attacking elderly or old people, which is very similar to the disease of childhood in method of onset, symptoms, duration, etc. Rheumatism is less frequently an antecedent, and organic heart disease is not so common. Cases by Roger,⁴ Russel,⁵ Saundby,⁶ Ferguson,⁷ Graves,⁸ and Gauthier⁹ are cited.

¹ Cyclopædia of the Practice of Medicine, vol. xiv. p. 423.

² Medical and Surgical Reporter, April 13, 1872.

³ Brain, April, 1888, p. 134.

⁴ Gazette des Hôp., 1854, xxvii. 559.

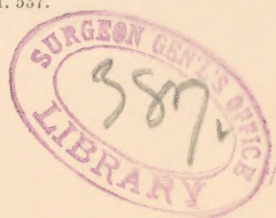
⁵ Medical Times and Gazette, 1879, ii. 447.

⁶ Lancet, 1884, ii. 948.

⁷ Lancet, 1885, ii. 92.

⁸ Clinical Medicine, i. 537.

⁹ Journ. de Méd. et Chir. Prat., 1881, 411, 66.



The *second* group comprises those cases dependent upon a coarse sub-cortical lesion. They often follow, but sometimes precede, hemiplegia. The lesion is usually a patch of softening. Hemichoreics are the chief class in this group.

The *third*—and the class I wish especially to consider—comprises those cases of the disease first appearing in adult age and continuing for years—usually until death. Members of the same family are affected, and direct hereditary transmission is easily traceable. Huntingdon is accredited with the first description of such cases in 1872. Mention is made of reports of cases by Ewald,¹ Peretti,² King,³ and others.⁴ The case which I shall hereafter record belongs to this class.

The *fourth* group, as it concerns us but little, I will not consider, except to say that it comprises all the remaining cases of chorea in the adult and in the old which cannot be placed in any of the three preceding classes.

Herringham's classification, while it can be of little scientific use, in view of our present scanty knowledge of the pathology of chorea in the adult, yet is convenient, and serves a practical purpose. Hammond,⁵ in his vast experience, has seen but four choreics in whom the disease began after thirty, and in none of them are we told there was a family history of chorea. With few exceptions, all the literature on the subject has sprung up since the date of the sixth edition of Hammond's book (1876).

Herringham, in his critical digest, made as late as last year, speaks of hereditary chorea as making the smallest of the four classes into which, we have seen, he divides chorea of the adult and of old age. A brief reference to reports of some cases which have come to my notice may, I trust, prove instructive.

Wharton Sinkler,⁶ in discussing the subject, makes reference to Huntingdon's report of hereditary chorea as being the first. Clarence King,⁷ in a valuable contribution to the subject, shows that mention was made of the hereditary chorea by C. O. Waters,⁸ of Franklin, N. Y., as early as 1841, while Irving W. Lyon's⁹ paper also antedates that of Huntingdon. In these cases, insanity and a tendency to suicide were common. I am unable to find reports of any cases between the years 1872, the

¹ Zeitschr. für klin. Med., vii., Supplement H, p. 51.

² Berliner klin. Wochenschr., 1885, No. 52.

³ New York Medical Journal, 1885, i. 468.

⁴ In a second critical digest (Brain, October, 1888, p. 415) Herringham gives a more extended consideration to this class of cases.

⁵ Diseases of the Nervous System, p. 715, 6th edition.

⁶ System of Medicine, Pepper, vol. v. p. 440.

⁷ Medical News, July 13, 1889.

⁸ Dunglison's Practice of Medicine, 3d edition.

⁹ American Medical Times, December 19, 1863.

date of Huntingdon's monograph, and 1881, when M. D. McLeod¹ makes an interesting contribution to the subject. He relates the cases of two sisters, admitted to the Cumberland and Westmoreland Asylum, Carlisle. These women had two brothers who were choreic. Their father also had had the "shaking disease" for many years before his death.

CASE I.—Isabella S., aged sixty-two, married, housewife, "peculiar" in disposition, admitted December 5, 1876, having been choreic for two years previous to that date. All muscles affected equally, apparently. Cessation of movements during sleep; mentally, irritable and childish. Physical health good. Short time before death twitchings became less frequent and intense on right than left side. Mental and physical powers gradually failed. Died August 19, 1880.

Autopsy.—Under dura of vertex and sides of left hemisphere there was a cyst lined with thick and dark-colored false membrane. The cyst extended over an area of brain comprising the posterior part of the frontal and whole of the parietal lobes of the left side; convolutions under this growth were depressed and flattened. Large gliomatous bodies on the choroid plexuses.

CASE II.—May B., widow, aged seventy-two, admitted September, 1879, sister of Isabella S. (Case I.). Chorea of two years' standing. Twitchings well-marked and general. No movements during sleep; great mental stupor. Paralysis of both legs. Died January 19, 1880.

Autopsy.—Several tumors in the texture of the dura mater; largest of them, about the size of a chestnut, was located in the upper part of the left hemisphere. Clustered around it were a number of small tumors, about the size of peas. They produced a distinct depression at the roots of the first and second frontal convolutions, and in the upper part of the ascending frontal and parietal convolutions, thinning, but not entirely destroying, the underlying cortical matter. The tumors were of hard, scirrhous nature; arteries atheromatous.

In several respects these cases differ from so-called "hereditary chorea," but, on the other hand, they show a far greater resemblance to that affection than they do to any other disease. If it were doubted that the father had chorea, would not the fact of the existence of the disease in the two brothers be strong presumptive evidence of heredity as a prominent etiological factor? I assume that they properly come under the head of "hereditary chorea." It seems remarkable, indeed, that in both cases such gross cortical irritants or destructive agents should have been found *post-mortem*. If these cases had been under medical observation in their beginnings, perhaps, too, it would have been noted in each case that the movements began on the side opposite to that of the brain upon which the growth was situated. Is it not likely that the disease affecting the two brothers of these women is also dependent upon a coarse lesion of the brain or its envelopes?

Ewald² records the instance of a family in which nine persons, representing three generations, were choreic. He also speaks of a mother, who, with her two daughters, was affected—the disease beginning in all cases after the age of thirty years. King has three times recorded cases

¹ Journal of Mental Science, July, 1881.

² Zeitschr. für klin. Med., vii., Supplement H, p. 51.

of hereditary chorea. In his first report in 1885¹ he describes the disease which affected many members of one family in three generations. More or less insanity was noted in all these cases. December, 1886, he reports another case.² In July of the present year he makes a third report.³ Mrs. C., who died of chorea at an advanced age (having been affected many years), had four children, only one of whom, Abel C., was affected with the disease; the other three children escaped, and none of their children developed chorea. Abel C. had six children, all of whom lived to have families. Two of his children, a son and a daughter, became affected. The son now is the father of two children who are said to show signs of the disease.

These cases would seem to be a strong corroboration of Huntingdon's law, that if one member of a family escape the affection the hereditary influence is stamped out in his branch of the family.

West gives a good description of a family of choreics. Huber⁴ records a series of cases: A man, aged thirty-eight years, had been choreic eight years; movements exaggerated and universal. His sister, father, two paternal uncles and an aunt, and paternal grandfather and great-grandfather were affected.

Zacher⁵ records a case of a man, aged forty-five years, who had developed the disease four years previously. He was committed to a hospital for the insane with the history that he had often abused his wife since the advent of his affection, and was subject to violent attacks of rage and excitement. Physical health was good; all muscles affected, and patient walked with great difficulty; speech was also much affected. By a strong effort of the will he was able to write his name. Entire quiet of muscles during sleep. The patient's grandfather was choreic, and transmitted the disease to one daughter and one son; his remaining child, a son, did not develop the affection. The daughter had seven children, three of whom had developed chorea—one of the three being the patient. In all the cases where a reliable history was obtainable it was found that the disease began between the ages of forty and forty-two years.

Hoffman⁶ reports four cases under his observation—two brothers and a sister of one family, and a female cousin. They all developed the disease between the ages of thirty and forty years, except one who has had epilepsy since the age of two or three years, and who became choreic while yet attending school. The disease was shown to have run through four generations, attacking, in all, thirteen individuals.

¹ New York Medical Journal, 1885, i. 468.

² Medical Press of Western New York.

³ Medical News, July 13, 1889.

⁴ Virchow's Archiv, Bd. cviii., H. 2, 267, 268.

⁵ Neurol. Centralblatt, January 15, 1888.

⁶ Virchow's Archiv, cxi. 3, 513.

None of the children of the fifth generation, all of whom are under twenty-four years of age, have yet developed the disease.

Lannois¹ reports cases of a brother and sister who inherited chorea, and adds a summary of cases already published.

Ducellier² gives a lucid description of a typical case.

Dr. Gershom H. Hill, Superintendent of the Iowa Hospital for the Insane, furnishes me with the following notes of a case of chorea under his care in that institution :

Mary S., aged fifty-five, married, but childless. Choreia began insidiously twelve years ago, and has steadily increased until she is now hardly able to walk, feed, or dress herself. Her mind has been affected for about two years. An uncle is choreic, and a brother is seriously threatened with the same disease.

From the hospital archives Dr. Hill has also given me notes of two additional cases :

Male patient: a case of dementia, admitted fifteen years ago, with history of having had chorea for a long time. He had wandered from home in the winter season, and as the result of this suffered from frozen feet. His feet were amputated, and he died a few days afterward from the effects of the operation.

Two years ago a brother of this man, aged forty, was admitted to the hospital, having been affected with chorea six years. He died of exhaustion, after a residence of twenty-one months in the institution. The history states that a sister of these two patients has been insane, but does not inform us as to whether or not she had chorea.

For notes of the next case—one in the State Lunatic Asylum, Utica, N. Y.—I am indebted to the Superintendent, Dr. G. Adler Blumer :

Female, aged fifty-six, unmarried. Insanity began 1884, and was coincident with the appearance of chorea. On admission her mental condition was that of melancholia. She has since become somewhat demented. Choreia has gradually increased. Her mother and a maternal cousin had chorea. In the case of her mother chorea came on during the last years of life (a number of years after the birth of the patient). She has no heart trouble and never has had rheumatism.

I will now give the principal points of interest I was able to gather in a case under my care in this hospital :

Hannah B., aged forty-seven years, widow, mother of four children, ranging in ages from nine to twenty years, was admitted to this hospital July 9, 1888, on the usual certificates of insanity signed by two qualified practitioners.

Statement which accompanied the certificate recites that she had been "weak-minded," and affected with St. Vitus's dance for the past six or

¹ Revue de Médecine, August 10, 1888.

² L'Encéphale, December, 1888.

eight years; she had threatened to poison some of her neighbors, and also to take her own life; and on one occasion actually did attempt suicide by drowning. Her normal disposition, which was quite amiable, has become perverted, and she is now peevish, irritable, and querulous, prone to sudden and violent fits of anger. Often, too, she is very despondent, and during such periods she has threatened or attempted violence to herself and others.

Condition on admission.—Rather spare frame, a little above the average height; dark eyes and hair, dark complexion. Facial lines deep; expression somewhat melancholic. Presents the appearance of one enjoying fair physical health. No abnormalities of organs of respiration, circulation, digestion, or excretion are noted; appetite good. Choreic movements noted in face, neck, arms, legs, and trunk. When she is alone, sitting or lying down, there is great diminution in the intensity of the movements. On one occasion like this the muscles of the neck were the only ones which appeared to be disturbed. Mental excitement or physical exertion causes great exacerbation of her symptoms. The effort necessary in talking causes marked exaggeration of the movements; but they can, in a large measure, be controlled by an effort of the will. All movements are arrested during sleep.

Present condition.—Physical condition is rather improved upon that noted upon her admission. She is usually melancholic and disposed to rail at friends and authorities by whom she was placed here. It is not known that she has made any threats of suicide or homicide since her advent to the hospital; but she is restless, irritable and quarrelsome, and often provokes to anger the other patients in the ward. Effort of talking always greatly increases the movements. An attempt which she made to write her name proved a failure—the first letter being the only one which was legible. It is only by considerable effort she walks; several times has fallen.

Her appetite is apparently insatiable. She often provokes and angers the other patients in the dining-room by appropriating to her use food and drink intended for others. Although her liberal allowance of food has been increased, her propensity for “foraging” among her neighbors is still apparently unabated.

Family history.—Our patient, the fourth child of her parents, has nine brothers and sisters—all of adult age, and four of whom are choreic—two sisters and two brothers. The father of these children, two paternal aunts, and two paternal uncles had the “shaking disease” for many years. Mrs. B.’s grandfather was also undoubtedly choreic.

One of Mrs. B.’s children had an attack of chorea at the age of eight, which lasted about two months. The child entirely recovered from this, which appears to have run the ordinary course of chorea of childhood. Mrs. B.’s uncles and aunts who were choreic all married and had children, but I was unable to ascertain that any of them inherited the family malady. None of the children of her four choreic brothers and sisters have developed the disease, but they are all under twenty-five years of age. To sum up: In one family we have eleven cases of chorea in three generations, or twelve cases in four generations if we include the case of Mrs. B.’s child, who, we have seen, developed the ordinary chorea of childhood.

This history I have verified from several sources, and as far as it goes I believe it to be entirely correct. The patient herself was able to give

me a good deal of information, which was afterward corroborated by statements of a relative who visited her. The family is now scattered, and it is likely that there are members who are affected other than those here recorded.

I was unable to learn definitely the time of life at which the affection appeared in each of these cases, but in the instance of Mrs. B. it was at the age of forty, and I am inclined to believe that all or most of the others developed the disease at or about that time of life. In no case has a recovery ensued, but, on the other hand, they all tend to a progressive exaggeration of the disease. Various degrees of mental symptoms developed in most cases, from an undue irritability of temper to the degree represented by our patient, who is the only member of the family who has ever been in an insane asylum. The neurosis, however, does not appear to be incompatible with a fair degree of physical health, some of the members of the family having had the disease for many years prior to their deaths—in one case as long as twenty years.

REMARKS.—An interesting and unique feature of the history of this family is the chorea which Mrs. B.'s child developed at the age of eight, and which apparently ran the ordinary course of the disease common in childhood. I am unable to find an instance similar to this in the writings of any of the authorities whom I have quoted in this paper. Would not a case like this be an argument against the views of those writers who maintain that chorea of old age, and especially "hereditary chorea" or "Huntingdon's disease," is essentially different from the affection seen in childhood—not because it is dependent upon a different morbid process, but because of the difference in clinical histories and in the durations of the two troubles?

The *Lancet* of August 17, 1888, in an editorial comment, says:

"Ordinary chorea is sometimes hereditary, but the disease called 'hereditary chorea' is, in many respects, different from the chorea of childhood. The distinction . . . rests . . . rather upon the characteristics, and chiefly upon the time of life at which the diseases become prevalent, and upon their observed progress. Child chorea ever tends to get better . . . ; whereas, hereditary chorea once established never ceases to exist. Hereditary chorea is frequently, child chorea is rarely, attended with marked mental chorea, if such a phrase may be allowed to signify the jerky, weak intellectual and moral actions of the sufferers."

This concise *résumé* is accurate in every statement, and while I readily subscribe to all it contains, I cannot see that there is adduced sufficient reason to warrant the invention of a "brand new" disease with half a dozen synonymous names.

PATHOLOGY.—I assume to be correct the theory, as promulgated by Hughlings-Jackson and Broadbent, that chorea of childhood is dependent upon certain changes or disturbances in the cerebrum. But the fact that small cerebral emboli have been found *post-mortem* in children who have had rheumatism, with endocarditis, does not prove that chorea of childhood is solely dependent upon this cause. Many cases have been

recorded in which the hearts were healthy and where diligent search failed to discover any emboli in the brain.

In cases where emboli are found in the corpus striatum, or other parts of the brain, it seems a very plausible example of cause and effect. But how are we to account for those cases where a critical examination of the brain gives a negative result, or, at most, shows some excess of serum, or a slight meningitis?

Koch's theory is very inviting. He says there exists a choreic virus or blood dyscrasia, which in some cases produces chorea, in others rheumatism, and again in others both these affections.

The pathological records of cases of "hereditary chorea" are so meagre that we have but little opportunity to study the subject.

McLeod's report of the cases of two sisters affected with "hereditary chorea," in both of which gross lesions producing cortical pressure were found *post-mortem*, seems to me to afford food for much thought. Might not the father of these women have also suffered from a similar lesion, and transmitted to his children an hereditary tendency to intracranial growths? Perhaps it was syphilis. Certainly his nerve functions were weak. I am inclined to think that records of autopsies which will be made in the future will show that the affection is not dependent upon any one well-defined morbid process, but, like epilepsy, upon a variety of changes in the brain or its envelopes, all producing essentially the same motor disturbance. I believe, further, that the only difference between chorea of childhood and that of adult age and old age (including the hereditary variety), will be shown to be simply a difference in the length of time the causes operate, the effect on motor cells or tracts and the symptoms produced thereby being practically the same. A choreic virus, a blood dyscrasia, or a number of minute emboli may soon disappear. The existence of these causes is too transient to produce an injury or "habit" from which the motor apparatus of the brain cannot recover. On the other hand, in adult age all diseases have a greater tendency to become chronic than they do in childhood. Gross brain lesions, such as those in McLeod's two cases, are a permanent and lasting cause; so is a clot—organized or broken down. Post-apoplectic choreics must be accounted for from this last cause. In some cases delicate chemical changes, or a fine general sclerosis, may be the permanent excitant or irritant. Here it might be difficult or impossible to demonstrate the changes, and then the disease would be said to be "functional."

Congenital chorea and cases where the disease began in childhood and continued many years, may be considered as arguments favoring the views I have here ventured to express.

Cases in which chorea and epilepsy exist in the same individual at the

same time, or in which the one affection has followed or supplanted the other, are also instructive as bearing upon this subject.

Dr. C. B. Mayberry has under his care in this hospital a case of this kind:

A boy, aged sixteen years, has chorea alternating with epilepsy. He never has a fit during the time of the existence of the choreic movements. The choreic movements last from one to three weeks, and after an interval of about the same length of time, they will again become manifest. During this interval, epileptic fits are of frequent occurrence—ten or fifteen in a day having been noted. Fowler's solution has a marked effect in controlling both of these manifestations of motor explosions, as has also bromide, though in a much less degree.

The diseases of the motor apparatus of the brain are not always so clearly defined and distinct from each other, either in morbid anatomy, symptoms, or course, as some authorities would lead us to believe; but on the contrary, are often so associated with each other that a precise diagnosis is difficult or impossible.

Would it not, then, be better to group and classify these diseases, and thus work in a broader and more comprehensive way, rather than to make artificial distinctions, with the consequent multiplication of names and synonyms? Let us not, at present, make two diseases out of chorea, but simply call each a *variety* of the same affection—at least until we have added much more to our knowledge of this interesting neurosis.

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